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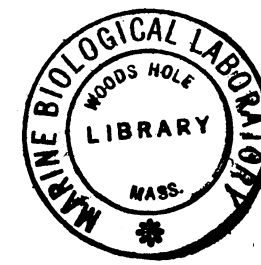
Total intracardiac repair of the adult cyanotic tetralogy of Fallot:

Clinical experience and late follow-up



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Summary: Twenty-seven adult patients with the cyanotic type of tetralogy of Fallot had total intracardiac repair during the past five years at the Toronto General Hospital. Twenty-one of 27 patients (77%) had had a previous shunting operation performed five to 21 years prior to the total intracardiac repair. Seventeen of 21 (81%) of these anastomoses were patent at the time of the total intracardiac repair. The hospital mortality was 11% and was attributable to hemorrhage – a well-recognized problem in adult tetralogy, due to excessive collateral circulation. All 24 surviving patients followed up from six months to five years showed marked clinical improvement, with disappearance of the cyanosis and reduction in acne and clubbing. Permanent heart block has not occurred. Late hemodynamic studies showed a significant persisting shunt in two out of 16 or 12%.

Since Lillehei's first description 15 years ago of total intracardiac repair for the tetralogy of Fallot,[1] numerous reports of the early and late results in mixed series of patients have become available.[2-12] Few of these have concentrated on the truly cyanotic form of this complex anomaly in the group over 16 years of age.[13, 14]

Is the surgical risk comparable in the child and adult? What is the duration of the long-term effect of previous palliative operations? Are the late results of intracardiac repair in the adult as satisfactory as in childhood? To answer these questions this review of 27 consecutive adult patients operated upon at the Toronto General Hospital was undertaken.

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Materials and methods (Table I)

1. Clinical material

Over the past five years, 27 patients between the ages of 16 and 48 years have undergone total intracardiac repair for tetralogy of Fallot. Five were under 20 and four were over 30 years of age. The majority (18) presented in the third decade. Most worked or attended school, but led socially and physically restricted lives because of fatigue and cyanosis at rest or on exercise. Clubbing was demonstrable in 20, and about half the patients had hemoglobin levels over 20 g. per 100 ml. at the time of intracardiac repair.

2. Previous palliative operations

Twenty-one patients (77%) underwent some form of palliative operation between the ages of 2 and 15 years. One had a Brock procedure[15] and one a functioning Pott's[16] anastomosis for 16 years. Single Blalock-Taussig[17] shunts had been performed in 15 patients, and four had had bilateral Blalock procedures. Seventeen of these 21 shunts (81%) performed eight to 20 years earlier were still functioning at the time of total correction. Of the four bilateral Blalock procedures, the first operation had been undertaken between the ages of 2 and 5 years and one functioned for 21 years. The second operation was carried out between 8 and 13 years of age, and the shunt remained open in all patients for 8 to 14 years.

3. Anatomical findings

All patients had large ventricular septal defects below the level of the crista supraventricularis, measuring 1.5 to 2.5 cm. in diameter. In one patient the defect extended to the annulus between the pulmonary and aortic valves. In another there was an associated "Swiss cheese" defect near the apex of the septum. Aortic insufficiency due to prolapse of the non-coronary cusp into the ventricular septal defect complicated a third.

TABLE I
Total intracardiac correction of cyanotic type of tetralogy of Fallot

Age distribution (years)	*No. of patients*	
16-20	5	
21-30	18	
31-50	4	
Previous palliative operations		
No previous palliation	6	
Single Blalock anastomosis	15	
Bilateral Blalock anastomosis	4	
Pott's anastomosis	1	
Brock procedure	1	
Patency of previous palliative operations		
With functioning Blalock anastomosis	16	
With functioning Pott's anastomosis	1	
Total functioning anastomoses	17	(81%)
Total number of previous anastomoses	21	
Anatomical findings at operation		
Ventricular septal defect	27	
Fibromuscular infundibular stenosis	17	
Diffuse tubular right ventricular hypertrophy	10	
Pulmonary valvular stenosis	9	
Associated aortic insufficiency	1	

The level of obstruction in the right ventricular outflow tract and the size of the pulmonary valve annulus and pulmonary artery proved important. Seventeen patients had localized fibromuscular infundibular stenosis with a distal thin-walled infundibular chamber. Two of them had additional valvular stenosis, but all had a pulmonary annulus and distal pulmonary artery greater than 15 mm. in diameter.

The other 10 patients had diffuse, tubular hypertrophy right up to the pulmonary annulus with no infundibular chamber. In three the ring and distal artery measured 8 to 12 mm. in diameter. The other seven patients had varying degrees of valvular stenosis, with the annulus and distal artery measuring 10 to 18 mm. in diameter. All patients had developed severe secondary hypertrophy of the body of the right ventricle below the level of obstruction.

4. Operative technique

Through a median sternotomy, the patency of any previous shunting procedure was confirmed by palpation. The Pott's anastomosis was closed from inside the left pulmonary artery, utilizing deep hypothermia and temporary interruption of cardiac bypass. All Blalock shunts were isolated prior to bypass by dissection along the aortic arch or innominate artery to isolate the appropriate subclavian vessel.[2] This technique avoided complications resulting from dissection in the area of dense adhesions about the anastomosis itself.

Total cardiopulmonary bypass was instituted once the anatomy had been confirmed. Over the past 1½ years, low volume prime, a disposable bubble oxygenator and normothermia have been utilized. Operation during this time has been simplified by anoxic normothermic cardiac arrest.

Through a transverse ventriculotomy,[18] avoiding any major or aberrant coronary arteries, an extensive ventricular resection[19] was begun, concentrating on the anterior and lateral margins of the outflow tract. Muscle excision proved simplest in those patients with an isolated infundibular chamber. In those with diffuse tubular hypertrophy it was always extended to the level of the valve ring. Portions of the fibrous thickening were removed from the posterior or cristal portion of the outflow tract, carefully avoiding perforation of the aortic sinus of Valsalva. Areas of the septum were also excised but superficially, to minimize laceration of the septal artery or perforation of the septum. Extensive resection of muscle from the body of the right ventricle, while avoiding the origin of papillary muscles, completed the creation of an adequate outflow tract.

When exposure of the pulmonary valve from below proved difficult, it was exposed from above through a pulmonary arteriotomy. Commissural incision was accomplished by direct vision in all instances of valvular stenosis. In those patients with narrow pulmonary valve annulus with or without stenosis, an incision was carried into the valve ring and followed by digital dilatation to produce a certain "give and stretching" of the ring. In no case was a pulmonary outflow patch inserted despite the small size of the pulmonary valve annulus, nor were residual outflow tract gradients measured on the operating table.

Closure of the ventricular septal defect then followed, using a woven Teflon patch and interrupted pledgetted mattress sutures. Care was taken to incorporate the annulus of the septal leaflet of the tricuspid valve and only the right side of the ventricular septum up to the papillary muscle of the conus to avoid the conduction bundle.

One patient underwent concomitant aortic valve replacement because of associated aortic insufficiency prior to operation.

Results (Table II)

A. Mortality

There were three hospital deaths. Postoperative hemorrhage was a major complicating factor in all three. Earlier exploration in two might have changed the outcome. The third, a severely cyanotic patient, developed a generalized bleeding tendency with coagulation abnormalities that proved impossible to correct. It was confirmed at postmortem examination that in all three patients the cardiac repair had been adequate.

TABLE II
Mortality and complications

	No. of patients	*%*
Total number of patients	27	100
Mortality	3	11
Postoperative hemorrhage	13	48
Permanent heart block	0	0

B. Complications

(1) *Operative*

(a) *Hemorrhage* — The development of excessive collateral blood flow necessitated meticulous hemostatic technique with suture and cautery prior to and after cardiopulmonary bypass. Excessive bronchial flow obliterating the operative field during repair was alleviated by venting the left ventricle.

(b) *Heart block* — No patient developed heart block, but right ventricular epicardial electrodes were always inserted as a precautionary measure.

(2) *Postoperative*

(a) *Hemorrhage* — Postoperative bleeding proved a problem in nearly one-half of these patients, necessitating early re-exploration. In most cases an actively bleeding collateral vessel was found and controlled.

Some patients had demonstrable pre- and postoperative coagulation abnormalities, the most frequent of these being fibrinolysis. When demonstrated by postoperative studies, any such abnormality was managed by appropriate medication.

(b) *Respiratory insufficiency* — Many patients had low arterial oxygen tensions for three or four days after operation. Early in this series prolonged respiratory support, often with tracheostomy, was utilized to combat this arterial desaturation. It then became evident that pulmonary shunting of excessive collateral flow was responsible and was unresponsive to ventilatory support. This degree of arterial desaturation appeared to be well tolerated by patients accustomed to cyanotic heart disease. As a result tracheostomy has rarely been undertaken in the latter half of this series.

Late follow-up data (Tables III and IV)

Twenty-four patients were well, active and completely rehabilitated six months to five years after total intracardiac repair. None was cyanotic and all had marked reduction in clubbing and acne; these changes proved most important in enabling them to adopt an entirely different outlook on life. Most were on no medications except prophylactic antibiotics during dental care and intercurrent infections.

Six patients had no murmur when reassessed. Two had Grade II pansystolic murmurs over the left sternal border at the 4th interspace and the remainder Grade I to II systolic ejection murmurs over the right ventricular outflow tract. Three had diastolic murmurs thought to be due to pulmonary insufficiency. None had evidence of congestive cardiac failure. All had complete right bundle branch block. There was no instance of permanent complete heart block.

Sixteen of the 24 long-term survivors (66%) consented to hemodynamic studies six months to five years after intracardiac repair. Two patients were found to have significant (2:1) left to right shunts at the ventricular level due to leaks about the septal defect patch. Two have left to right shunts below 1.5:1 and the remainder have none.

Two patients have residual gradients across the pulmonary valve annulus of 40 to 50 mm. Hg. The remainder have insignificant residual gradients under 20 mm. Hg. Nine demonstrated low cardiac indices and 12 had mild elevations of right ventricular end-diastolic pressure. Five responded normally to exercise.

TABLE III
Clinical assessment of patients followed up for six months to five years

	No. of patients	%
Total number of patients seen	24	100
Disappearance of cyanosis	24	100
Reduction or loss of acne	24	100
Reduction or loss of clubbing	24	100
Complete right bundle branch block (ECG)	24	100
Complete heart block	0	0
Congestive heart failure	0	0
Heart murmur present	18	75

TABLE IV
Catheterization data after total correction of adult cyanotic tetralogy of Fallot (from six months to five years later)

	No. of patients	%
Total number of patients studied	16	100
Significant residual left to right shunt (2:1)	2	12
Small residual left to right shunt (1.5:1)	2	12
Significant gradient across pulmonary valve	2	12
Mild elevation of right ventricular end diastolic pressure	12	75
Low cardiac index	9	56

Discussion

Total intracardiac repair of tetralogy of Fallot entails a published operative mortality of 7 to 30%.[3, 5, 10, 11] Most of the published series, however, have included acyanotic as well as cyanotic patients, and children as well as adults.[3-6, 10-12]

We have presented our experience with 27 adult cyanotic patients with tetralogy of Fallot who underwent total intracardiac repair. Our mortality of 11% compares favorably with that of other series, especially when one considers that more than 50% of these adult cyanotic patients had hemoglobins of over 20 g. per 100 ml.[20] The mortality has been directly related to hemorrhage, a result of the extensive collateral blood flow.

It proved difficult to evaluate precisely the effectiveness and durability of earlier palliative procedures in most of these patients. Lack of detailed follow-up studies, development of collateral blood flow, and anatomical variations in the right ventricular outflow tract were some of the important variables.

Sixty-two per cent of the patients operated on had a functioning shunt procedure at the time of intracardiac repair. Despite this, cyanosis and polycythemia had progressed after the age of 20 years, especially in those who had undergone bilateral palliative operations. This deterioration may have been due to the fixed size of the shunt related to body growth or to progressive hypertrophy in the right ventricle increasing the degree of outflow obstruction. However, most of our patients had a small-sized left ventricle and small pulmonary arteries and annuli. Although shunt operations are an efficient means of palliation for these patients,[21] our experience contradicts previous statements that shunting procedures prepare the left ventricle for later correction.[22] On the other hand, our follow-up data reveal that after total repair the left ventricle is capable of carrying efficiently the workload of the heart. It is also of interest to note that 81% of these shunts had remained patent, one of them after 21 years. With or without a palliative shunt, all patients over the age of 20 years had developed a prolific collateral blood flow to the lung.

The least symptomatic patients in our series (2/3) were those with localized infundibular stenosis, an infundibular chamber and large pulmonary arteries and annuli. Some had undergone a previous shunting procedure but probably reached adulthood pri-

marily because of an attractive anatomical situation. Our late results suggest that these patients are the best candidates for total correction. They also make up the dominant population in published pediatric series.

The other one-third of our adult cyanotic patients were those with diffuse tubular hypertrophy which extended to the level of the pulmonary annulus. They tended to have a higher incidence of previous unilateral or bilateral palliative procedures and more severe cyanosis and disability.

The operative repair has been facilitated by the large adult heart and in the last 1½ years by anoxic cardiac arrest, even though extensive resection of secondary hypertrophy was necessary. This may have been important in avoiding heart block. Hemorrhage due to extensive collateral blood flow to the lungs can be a complicating factor during and after operation.

The late results have been gratifying. Congestive heart failure and complete heart block have not occurred, despite the fact that residual shunts and residual outflow gradients have occasionally been demonstrated by the hemodynamic studies. The majority of the outflow gradients have been markedly reduced. Similarly the problems of pulmonary valve insufficiency related to outflow tract patches have been avoided.

Conclusions

The total correction of adult cyanotic tetralogy of Fallot has always represented a serious decision, but this series would appear to demonstrate that it can be carried out with an acceptable low mortality rate with excellent clinical results. Adult tetralogies should be considered for assessment and surgery whenever they demonstrate a significant degree of deterioration.

Résumé

Réfection intracardiaque complète de la tétralogie de Fallot de forme cyanotique chez l'adulte: expérience clinique et post-observation

Au cours des cinq dernières années, nous avons procédé chez 27 adultes à la réparation intracardiaque totale de la tétralogie de Fallot de forme cyanotique à l'Hôpital Général de Toronto. Avant cette intervention, au cours d'une période antérieure variant de cinq à 21 ans, 21 de ces 27 malades (77%) avient déjà subi une dérivation. Au moment de l'intervention définitive, les anastomoses étaient perméables chez 17 de ces 21 malades (soit dans 81% des cas). La mortalité hospitalière a été de 11%: elle était généralement causée par l'hémorragie, complication fréquente de la tétralogie de l'adulte et résultant d'une circulation collatérale excessive. Les 21 survivants ont été suivis pendant une période variant de six mois à cinq ans. L'amélioration clinique a été remarquable: disparition de la cyanose et dimunition de l'acné et de l'hippocratisme. Nous n'avons observé aucun cas de bloc cardiaque permanent. Les dernières études de l'hémodynamique ont révélé une dérivation persistante chez deux malades sur 16, soit dans 12% des cas.

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